Specialty Conference

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Vascular Compression of the Upper Airway in Children

J. DEANE WALDMAN, MD:* *In the past 18 months we have seen ten children with upper airway obstruction due to vascular anomalies. In view of the rarity of this condition and the number of different specialties involved in the care of these children, we thought it would be instructive to have a symposium on the subject of vascular compression of the upper airway. Each of the participants has been asked specific questions within his area of expertise. It is hoped that these are the type of questions that a physician with such a patient would ask.*

Dr. Eli Meltzer is both a pediatrician and an allergist; he was asked "How do I recognize one of these children?" Dr. Ken Miller, pediatric radiologist for the Children's Hospital, was asked "What does radiology have to offer in the evaluation of such a child?" Pediatric pulmonologist Dr. Sung Park was saddled with a deceptively simple query: "What is the pathophysiology of obstruction of the upper airway?" As cardiac consultant, I have carried out the invasive studies in these patients and, therefore, my topic will be the differential diagnosis. Dr. John Lamberti is a cardiovascular surgeon with extensive experience in congenital anomalies; he will discuss surgical procedures employed in children with vascular compression of the upper airway.

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Clinical Data

ELI O. MELTZER, MD:† At the weekly CPC's at the University of Pennsylvania, there was an elderly clinician who regularly attended and regularly suggested kala-azar in the differential diagnosis of all conditions. Now even in Philadelphia, kala-azar is uncommon. Apparently, the physician wanted the students to note two things: (1) he was the only person who was right the one time that kala-azar was the correct diagnosis and (2) a not so subtle reminder of the old clinical saw "you have to keep a high index of suspicion." To continue our saga of medical homilies, there is the admonition that "when you hear hoofbeats, don't think *only* of horses."

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Today's conference is about some zebras, so to speak; about entities that may cause wheezing but are not asthma, about vascular obstruction of the upper airway. But these zebras are not all that rare, for we have seen ten cases here in San Diego within the last 18 months.

In reviewing the clinical histories of this group of children, certain points became evident. All were products of normal gestations and had normal birth weights; of note is the fact that all but one grew well (Table 1). Despite the presence of a fixed congenital obstruction, the clinical features were highly variable. The pattern was chronic, intermittent with mild-to-moderate (rather than severe) distress; wheezing or stridor and often both were noted, usually with a neonatal or early infant onset of symptoms (Table 1). Recurrent pneumonia was common. A history of swallowing difficulties, especially with solid foods, was occasionally a clue to a fixed, structural disorder of both the esophagus and trachea. In addition, some children were prone to recurrent aspiration pneumonitis.

Findings on physical examination frequently were unremarkable except for large airway noises, variable in nature. Indeed, other than locating the site of difficulty, physical examination was usually of little diagnostic help. Roentgenograms of the chest or bronchoscopy frequently pointed to the vascular system or at least to a discrete extrinsic compression of the upper airway.

The oldest child in our series (5 years old at presentation) had chronic cough and recurrent pneumonia. Wheezing had been reported frequently in the past. On pulmonary function studies, large and small airway obstruction was noted which did not change acutely when epinephrine was given or chronically with prednisone therapy. These results suggested a fixed, large airway disorder subsequently confirmed to be a vascular ring.

One final lesson should be derived from the clinical data. In pediatrics we are all trained to integrate abnormalities into a single disease entity because children very rarely have two separate but concomitant problems. In one of the two girls in our series (patient 1, Table 1) there was a strong family history of atopy; wheezing with and without infection, recurrent otitis, rhinitis and pneumonia were present, as well as both gastrointestinal and respiratory symptoms. She was intolerant of cow's milk, had eosinophilia and documented reaginic activity. In short, there was one very obvious cause of the recurrent respiratory distress: allergic hyperreactive small airway disease. Nonetheless, she *also* had a vascular ring; following surgical operation prominent clinical improvement was noted.

In conclusion, one must amend our old adage to say "When you hear hoofbeats, think of both horses and zebras, remember that they each have various species which can even run together."

Radiologic Evaluation

KENNETH E. MILLER, MD:* In evaluating possible vascular compromise of the airway, radiologists must evaluate three areas in stepwise fashion: (1) the great vessels on plain films, (2) the tracheal airway using magnification film technique[1,2] and (3) the esophagus on a barium swallow.

The Great Vessels

In plain films of a patient with obstructive airway disease, the presence of a right aortic arch suggests the possibility of vascular ring. In a normal child, the trachea is deviated slightly to

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TABLE 1.—*Clinical Characteristics of Children With Vascular Compression of the Upper Airway*

Number	*Diagnosis*	*Age at Presentation*	*Episodes of Pneumonia*	*Other Symptoms*
1. . .	Right arch/left ligamentum	4 m	9	Wheezing; rhinitis, otitis, swallowing difficulties
2. . .	Right arch/left ligamentum	1 w	1	Stridor; wheezing
3. . .	Right arch/left ligamentum	5 y	4	Cough; wheezing
4. . .	Right arch/diverticulum of Kommerell	8 m	1	Stridor; swallowing difficulties
5. . .	Innominate artery syndrome	3 w	0	Stridor
6. . .	Innominate artery syndrome	1 m	0	Stridor
7. . .	"Innominate artery syndrome"; tracheal stenosis	3w	4	Stridor; wheezing
8. . .	Ectopic thymus	2 w	2	Severe stridor
9. . .	Pulmonary artery sling	3 d	0	Stridor; wheezing
10. . .	Tetralogy of Fallot with absent pulmonary valve	1 d	2	Wheezing; failure to thrive

d = days; w = weeks; m = months; y = years

the right on the frontal roentgenogram; a soft tissue shadow to the *left* of the trachea just above the level of the bifurcation is the normal left arch. Tracheal deviation to the left is always abnormal[3] and the presence of a *right* paratracheal mass is the clue to a right arch (Figure 1A). In addition, usually a portion of the descending aorta can be seen behind the heart and adjacent to the vertebral bodies on well penetrated frontal films. Occasionally, the side of the aortic arch may be unclear, especially if the trachea is in the midline, surrounded by equal bilateral soft tissue masses; this may indicate the presence of a vascular ring with double arch (Figure 1B).

Magnification Films

High kilovoltage filtered magnification films[1,2] of the upper airway (Figure 1A,G,I) are taken routinely in children with symptoms of airway obstruction. This technique provides more reliable and accurate information with less radiation exposure than conventional films. These films are taken primarily to evaluate intrinsic causes of airway obstruction, such as epiglottitis, croup, foreign body, congenital web or stenosis, hemangiomas, goiter and the like; these conditions are much more common than vascular compromise. In our experience, anterior displacement and bowing of the trachea on lateral films of the chest (Figure 1F) are more reliable indicators of an aortic arch vascular abnormality than is indentation of the anterior surface of the trachea.

Fluoroscopy

Fluoroscopy has proved of limited help and can be carried out in conjunction with esophagography. One can obtain information about airway dynamics such as ballooning of the hypopharynx during inspiration—indicative of airway obstruction. Tracheomalacia and laryngomalacia can be evaluated with fluoroscopy[4] although both are difficult to confirm.

Indentation of the anterior surface of the trachea may indicate vascular ring, aberrant innominate artery or aberrant left common carotid artery. In addition, such an indentation is present in a surprisingly large number of asymptomatic infants (Figure 1G).[5,6] According to MacDonald

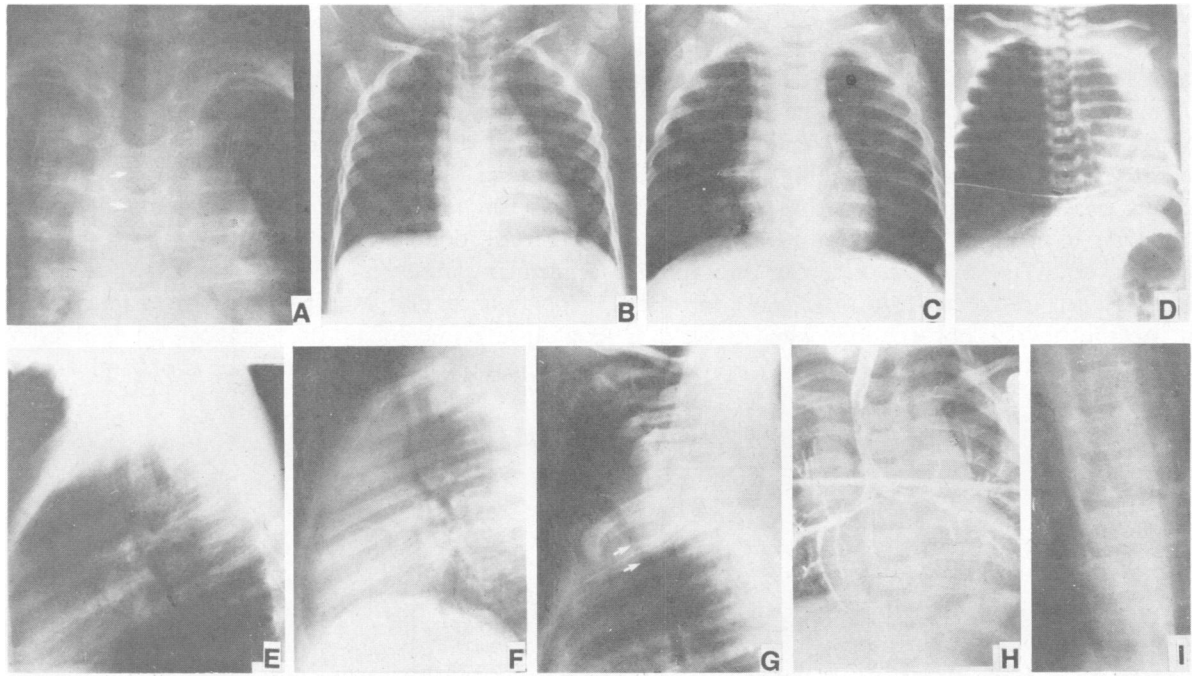

Figure 1.—Roentgenograms of the chest and upper airway. Situs of the aortic arch may be found on the right **A,** easily visible in high kilovoltage films of the upper airway or may be on both sides of the trachea **B,** as in double aortic arch. Pulmonary artery "sling" may produce bilateral air trapping **C,** or may affect only the right side **D,** and can be suggested on the plain chest film—lateral view **E,** by a mass between the trachea and the esophagus. Lateral chest films may show anterior bowing of the trachea **F,** cause by the aorta, diverticulum of Kommerell, or aberrant subclavian artery, or can demonstrate the *normal* airway narrowing seen as the trachea enters the lung. **G,** *Intrinsic* deformities of the trachea can simulate vascular compression: bronchogram **H,** shows generalized tracheobronchial narrowing in a patient initially thought to have "innominate artery syndrome"; high kilovoltage films in the same patient **I,** adequately show the tracheobronchial air column without instillation of lipiodol.

and Fearon[7] anterior indentation of the thoracic trachea should be evaluated by bronchoscopy, looking for a pulsating (presumably vascular) mass; in our experience, this has been frequently unrewarding.

Barium Swallow

The esophagogram provides essential information in the diagnosis of vascular anomalies. The following abnormalities should be sought: (1) oblique indentation made by aberrant right or left subclavian artery, (2) right or double aortic arch confirmation—the S sign on an esophagogram and (3) a mass between the trachea and esophagus—aberrant vessels such as a pulmonary vascular sling or nonvascular masses such as bronchogenic cyst, esophageal duplication cyst or hilar lymphadenopathy. Barium swallow also identifies tracheoesophageal fistula and gastroesophageal reflux, both of which can be associated with chronic aspiration producing reactive airway spasm which can simulate vascular compromise of the upper airway.

Esophagography in small children is potentially dangerous and should be carried out with oxygen, suction and intubation devices ready for use. Either a nurse or a member of the pediatric medical staff should be in attendance during the examination to feed and observe the child during fluoroscopy and to initiate any emergency procedures. We have avoided immobilization of the infants on rigid boards during these procedures preferring to maintain mobility of the child. When stressed, many infants with vascular ring abnormalities need to hyperextend their necks in order to breathe; this is not possible when a child is rigidly immobilized.

One rare situation can be misleading at esophagography; a right aortic arch may be associated with an aberrant left subclavian artery originating from an aneurysmal dilatation of the left descending aorta[8]: the so-called diverticulum of Kommerell (Figure 3B). This posterior mass is larger than the subclavian artery and may be misdiagnosed as a bronchial cyst, enlarged lymph node or esophageal tumor rather than a vascular mass. The diverticulum is frequently as large as the aorta and may displace both the trachea and esophagus anteriorly.

Simple aberrant right subclavian artery with normal left aortic arch is common (one in 200 autopsies) and usually asymptomatic and incidental. Most authorities regard it as an unlikely cause of feeding symptoms, the so-called dysphagia complex.

Mass Between Trachea and Esophagus

A mass located between the trachea and the esophagus (Figure 1E) causing upper airway obstruction can be seen on a lateral plain x-ray film of the chest in the pulmonary artery "sling." The aberrant course of the left pulmonary artery may compress the trachea above the bifurcation obstructing aeration of both lungs (Figure 1C) or, more commonly, may pass around the right mainstem bronchus causing a ball-valve phenomenon in the right lung producing unilateral emphysema (Figure 1D).

In summary, radiologists should look for the right or double aortic arch, and must exclude intrinsic causes of airway obstruction, evaluate the trachea and esophagus for aberrant vessels, and rule out reflux or tracheoesophageal communication as a cause of aspiration and airway

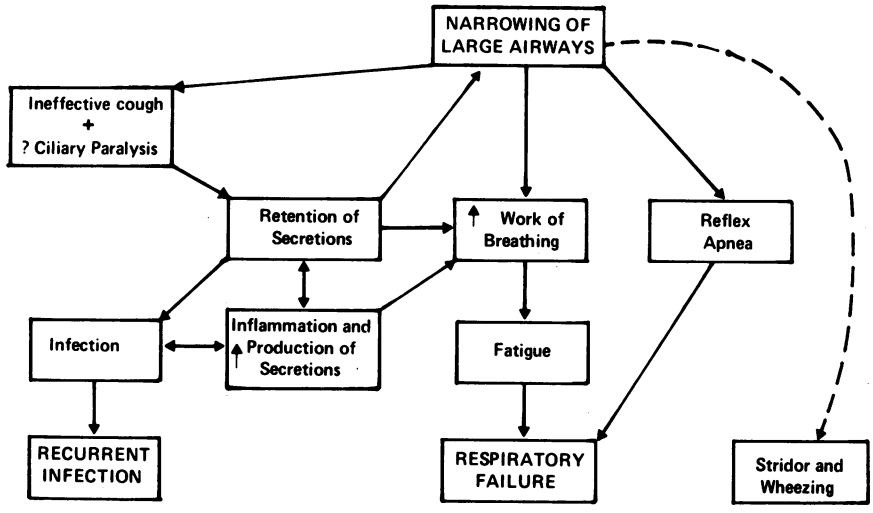

Figure 2.—Pathophysiology of narrowing of the upper airways.

compromise. We have been impressed with our inability to define anterior tracheal compromise in patients with proven vascular rings. We accept a right arch, aberrant (retroesophageal) left subclavian artery and clinical evidence of tracheal obstruction as prima facie evidence of a vascular ring. Close approximation of the trachea and esophagus during esophagography is a good indirect clue of a compressing vascular ring.

Pulmonary Pathophysiology

SUNG MIN PARK, MD:* Narrowing of the large airways can produce three results: (1) recurrent infection, (2) respiratory failure or (3) stridor and wheezing without clinical distress. The pathways from airway obstruction to the serious endpoints can be complex and are interrelated (Figure 2).

Respiratory Defense Mechanisms

Analyzing first the left-hand side of the diagram, we must briefly review the defense mechanisms available to the respiratory tract. There are four basic categories of respiratory defense: (1) mechanical, (2) immunological, (3) biocidal and (4) chemical. These mechanisms tend to work both in parallel and in series; therefore, one system can partially compensate for another. However, if one area is truly malfunctioning, compensation cannot continue indefinitely and the system breaks down, resulting in respiratory infection. For example, no matter how good the mechanical defense mechanism is, repeated respiratory infection occurs if the patient has immune deficiency disease. Another example is the patient with vascular obstruction of the upper airway, who tends to get recurrent respiratory infections although other defense components may be completely normal.

The Cough

Mechanical defenses include nasal air-conditioning, upper airway closure, mucociliary apparatus and cough. Although some authors believe that narrowing of the large airways itself can cause ciliary paralysis,[9] this remains unproved. The cough is one of the most efficient mechanisms to keep the lungs clear of secretions and foreign material. Let us review the dynamic events that occur in the large airways during the cough.[10] The tracheal lumen will reduce to a sixth of the original size with a cough producing a pronounced increase in the linear speed of the air stream. In patients with no obstruction, the reduction of tracheal lumen will leave enough space for the air stream to carry away the secretions. The cough can fail when one or more of the following conditions are present: (1) inadequate volume, (2) failure to trigger, (3) failure to achieve pressure and (4) airway collapse (failure to maintain patency) at the time of cough. With a vascular ring, the patient would have inadequate volume because of the stenosis, but would have normal trigger mechanism by the cough reflex. The pressure generated by the cough would also be insufficient because airway narrowing can restrict the inspiratory volume. Most important, however, the already narrowed airway would completely collapse, making the cough ineffective beyond the point of stenosis; thus, secretions will accumulate leading in turn to inflammation.

Recurrent Infection and Respiratory Failure

Returning to our general schema (Figure 2), we see that ineffective cough leads to retention of secretions, which in turn fosters infection, promotes inflammation and increases the work of breathing. All of these factors can lead to repeated clinical difficulties but infants are especially vulnerable. The infant airway caliber is, by nature, small and the cartilages of the airway are not as rigid as in older patients and therefore are easily compressed. Furthermore, the muscle reserve is considerably reduced in infants. The concept of muscle reserve is very important in understanding respiratory failure. Despite the presence of a pathologic condition in the lungs such as airway obstruction, as long as the patient has sufficient muscle reserve to overcome the added resistance, respiratory failure will not develop. On the other hand, if the patient does not have enough muscle or energy reserve to deal with the increased respiratory work induced by even mild airway obstruction, respiratory failure will occur quickly. Immaturity of the immune system in young infants puts them at a further risk.

Reflex Apnea

There is one other mechanism that can lead to acute respiratory failure: reflex apnea. MacDonald and Fearon[7] reported this occurrence in 50 percent of their patients with innominate artery syndrome in whom surgical operation was required. We have recently seen a patient who had an enormously enlarged left atrium compressing

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TABLE 2.—*Cardiovascular Causes for Compression of the Upper Airway*

AORTA
Double arch (patent or atretic limb)
Right arch
Retroesophageal aorta
Aberrant left subclavian artery
Left arch
Right ductus arteriosus
Aberrant brachiocephalic arteries
PULMONARY ARTERY
"Sling" (left pulmonary artery from right pulmonary artery)
Tetralogy of Fallot with absent pulmonary valve
LEFT ATRIUM
Large left-to-right shunt
Left atrioventricular valve regurgitation
Left atrioventricular valve stenosis

the large airways. His only complaint was of recurrent apneic spells. Therefore, either fatigue from increased work of breathing or reflex apnea can lead to respiratory failure.

Differential Diagnosis

J. DEANE WALDMAN, MD: Before understanding the subject under discussion, one must obtain a perspective on the total problem. There are many causes of upper airway obstruction: congenital defects such as choanal atresia and oral airway disorders, mass lesions whether neoplastic, infectious or foreign body, as well as conditions promoting edema or secretion can all induce obstruction to air movement. Vascular anomalies make

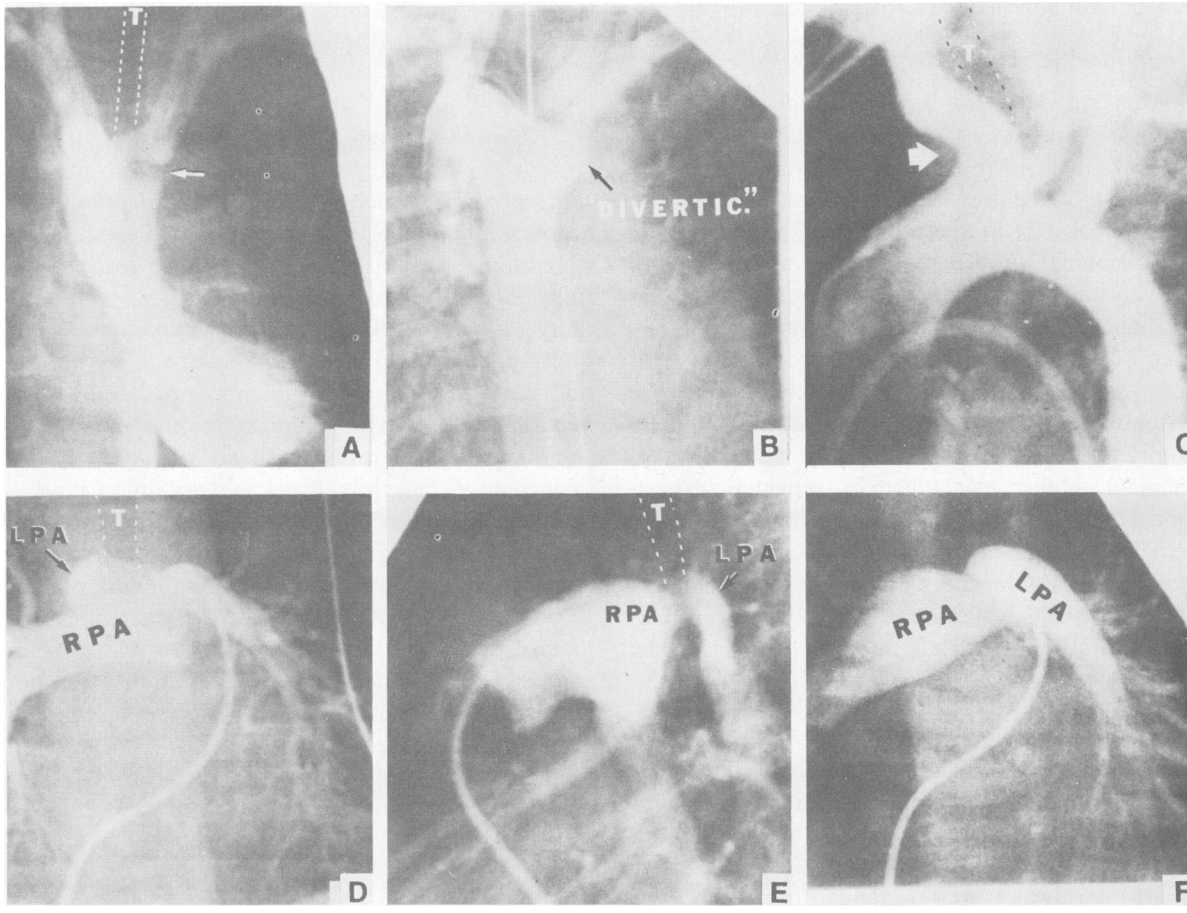

Figure 3.—Angiograms in vascular compression of the upper airway. A, Double aortic arch surrounding trachea (T); arrow indicates short atretic area in the ventral (left) aortic arch which can be severed surgically to disrupt the vascular ring. **B,** Oblique aortogram showing right aortic arch and diverticulum ("divertic") of Kommerell which is connected by the ligamentum arteriosum to the pulmonary artery thus completing the vascular ring. **C,** Lateral aortogram showing course of innominate artery—initially posterior (arrow) apparently compressing the trachea (T). **D,** Pulmonary arteriogram demonstrates origin of the left pulmonary artery (LPA) from the right pulmonary artery (RPA)—the pulmonary artery sling. **E,** Lateral view of pulmonary artery sling showing the trachea (T) encased by pulmonary arteries, the right (RPA) anteriorly and the left (LPA) posteriorly. **F,** Pulmonary arteriogram in patient with tetralogy of Fallot and absent pulmonary valve; there is massive dilatation of the right pulmonary artery (RPA) and moderate enlargement of the left branch (LPA).

up a small rare subgroup of conditions that produce airway obstruction.

There are three vascular structures that can compress the trachea or bronchus: the aorta, the pulmonary artery branches and the left atrium (Table 2).

The Aorta

There is a normal phase in the embryologic development of the aortic arch in which there are two aortas: ventral and dorsal. Usually, the dorsal aorta involutes leaving the ventral one to traverse the left mainstem bronchus, producing a left arch. Occasionally, only the dorsal aortic branch remains producing a right aortic arch.[11] Very rarely, both arches remain and fuse posteriorly to form the descending aorta. This condition—double aortic arch—always causes a vascular ring regardless of which side is dominant. While both branches around the trachea may be patent, it is possible to have a portion of one of the branches atretic (Figure 3A) with a fibrous cord connecting to the descending aorta and completing the vascular ring.[12]

It must be remembered that the ductus arteriosus or its descendant the ligamentum arteriosum essentially always lies to the left of the trachea. When the aortic arch is on the right and especially when part of the aorta is behind the esophagus, the components are present to form a vascular ring. The ring is formed by ascending and right-sided aorta, the retroesophageal aorta, the ligamentum arteriosum and the pulmonary artery. In many of these cases (Figure 3B) the retroesophageal aorta protrudes to the patient's left and has been called the diverticulum of Kommerell[8]; this structure is the landmark for the ductus arteriosus and is usually associated with a vascular ring.

An exceedingly rare condition has been described in which a vascular ring is formed by a left arch, retroesophageal aorta, and right-sided ductus arteriosus.[13] Review of published case reports suggests that this is either a complete mirror-image of the usual (right arch with left ligamentum) vascular ring or is a variant of double aortic arch with an atretic right (dorsal, anterior) limb.

Innominate Artery Syndrome

In the 1960's Berdon[5,14] and others wrote extensively about the "Innominate Artery Syndrome"; in this condition an aberrant course of the innominate artery is associated with anterior compression on the trachea and thus obstruction of the airway. Enjoying an immediate and enthusiastic vogue, the so-called innominate artery syndrome was extensively overdiagnosed and subsequently fell into disrepute. While the condition is clearly not as common as previously thought, it was diagnosed in three of our patients by angiography. In one child the condition was confirmed surgically, in one the symptomatology remained mild and no operation was carried out and the third patient (patient 7, Table 1) is worthy of detailed comment.

A 16-month-old infant with respiratory noises commencing at 3 weeks of age had four episodes of pulmonary infection within the first year of life. She slept in a supine position with her pillow behind her neck. Bronchoscopy carried out elsewhere suggested discrete anterior (but nonpulsatile) compression of the trachea. Aortic arch angiography (Figure 3C) showed an unusual course of the innominate artery from which also arose the left common carotid artery. *Innominate artery syndrome* was considered and the child was scheduled for arteriopexy. However, bronchoscopy was repeated preoperatively; a large segment of concentric stenosis of the lower trachea was found, shown by bronchography (Figure 1H). Thus, despite the apparent innominate artery syndrome, the patient's respiratory difficulties were intrinsic to the trachea rather than secondary to extrinsic (vascular) compression.

It is generally accepted that the thymus *never* causes compression of the upper airway. Our experience provided an exception to this dictum and emphasized the need to maintain a flexible attitude towards these patients. At operation in patient 8 (Table 1), an ectopic limb of the thymus was found behind the superior vena cava, connected to the main body of the thymus by a thick fibrous cord. Obstruction of the trachea was immediately relieved when the fibrous strand was severed.

Pulmonary Artery Branches

We now turn our attention from the aorta to the pulmonary artery (Table 2). When the left pulmonary artery arises from the right branch instead of the main trunk (Figure 3D) compression of either the trachea or the right mainstem bronchus can occur.[15,16] This is a condition in which, on a lateral x-ray of the chest, a mass is

seen *between* the trachea and esophagus (Figure 1E). A lateral angiogram (Figure 3E) dramatically shows how the trachea is narrowed posteriorly by the aberrant course of the left pulmonary artery coming from the right side. Note the tracheal air column "enveloped" by pulmonary arteries.

Tetralogy of Fallot with absent pulmonary valve syndrome is another rare cause of upper airway obstruction which we have seen recently. The unusual hemodynamics causes massive dilatation of the branch pulmonary artery,[17] usually the right (Figures 1D and 3F). Progressive enlargement and exaggerated pulsations of the pulmonary artery cause upper airway obstruction limited to the right side.

Left Atrium

In addition to the aorta and the pulmonary artery, the third vascular structure which can cause upper airway obstruction is the left atrium. Either left-to-right shunts or malfunction of the mitral valve can cause enlargement of the left atrium. Normally the left atrium is located just beneath the left mainstem bronchus; consequently dilatation of the left atrium presses up from below causing obstruction of the left bronchus and unilateral airway obstruction on the left.

Cardiac catheterization studies were done in each patient. Controversy does exist regarding the need for angiograms in all cases.[12] Techniques developed here[18] for aortic arch studies using the *venous* approach have stimulated angiographic studies because the dangers of retrograde arterial catheterization can usually be avoided. We feel strongly that no simple rule can be applied to all cases; individual determinations of proper diagnostic studies is the best approach.

Surgical Therapy

JOHN J. LAMBERTI, MD:* The surgical relief of upper airway obstruction caused by abnormalities of the aortic arch or its branches is generally straightforward. Before describing the surgical approach, I would like to comment on the diagnostic evaluation. Many surgeons believe that routine angiography or bronchoscopy is unnecessary in patients with an obvious vascular ring: stridor and roentgenographic evidence of tracheal compression in association with a diagnostic barium swallow are considered sufficient evidence for surgical therapy. Although angiography is of very little risk with present techniques, an aortogram is not required in all cases. In fact, in some instances, aortography may not clearly delineate the cause of airway narrowing. Nonetheless, angiography is indicated whenever the diagnosis is in question; oblique views may be helpful and several biplane angiograms may be necessary.

Bronchoscopy in infants—once a relatively high-risk procedure—has been well served by recent advances in technique, especially use of flexible fiberoptic bronchoscopes. When anomalous origin of the innominate artery is suspected as a cause of tracheal compression, bronchoscopic confirmation of a discrete anterior pulsatile tracheal compression is required before surgical intervention. However, in conditions such as double aortic arch, or right aortic arch with left ligamentum arteriosum, bronchoscopy is rarely indicated.

There have been relatively few innovations in the treatment of aortic arch anomalies since the first operation for this condition was described in 1945.[19] Double aortic arch or right aortic arch with aberrant left subclavian artery and vascular ring are all approached through a left thoracotomy which allows intraoperative delineation of the exact relationships. In 1953 Robert Gross summarized his experience with the surgical treatment of aortic arch anomalies[20] and, more than 25 years later, his recommendations continue to be valid. In cases of double aortic arch, the surgeon must divide the smaller or atretic aortic arch (usually the anterior one) in order to maintain adequate blood flow to the systemic circulation. The operation consists of division of the ligamentum arteriosum followed by division of the aortic arch at its narrowest point. Usually this will be between the left subclavian artery and the descending aorta. On occasion it may be technically easier and anatomically more appropriate to divide the arch between the left carotid and left subclavian artery. In such cases, the left subclavian artery becomes the distal-most branch of the right aortic arch system. In most cases, when the vascular ring is divided, immediate release of vascular compression is noted by the surgeon and the anesthesiologist may note that airway resistance has decreased. In cases of right aortic arch with aberrant origin of the left subclavian artery and left ligamentum arteriosum, there may be posterior compression of the esophagus as well

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as airway compression. Early descriptions of the surgical treatment of this entity indicated that the ligamentum arteriosum and the left subclavian artery should be divided. In such cases the vertebral artery should be ligated to prevent the subclavian steal syndrome. At the University of Chicago we reimplanted the left subclavian artery into the left carotid artery at the time of operation for this type of vascular ring on two occasions. This approach preserves blood flow to the left upper extremity as well as dividing the vascular ring and relieving posterior compression of the esophagus.

When the tracheal compression is caused by an anomalous innominate artery, the operation is best carried out by means of a median sternotomy or right lateral thoracotomy. The procedure involves anterior fixation of the innominate artery to the sternum. As previously indicated, innominate artery compression of the trachea is a rare cause of vascular compression and intraoperative bronchoscopy is an integral part of the therapy for this subset of patients.

Pulmonary artery sling has been surgically treated for many years and most early reports indicated satisfactory relief of tracheal compression. The left pulmonary artery is divided at its origin and transplanted to its normal location.[15] However, adequate long-term patency of the left pulmonary artery-to-main pulmonary artery anastomosis has been difficult to achieve.[16] This operation can be done through a left thoracotomy. We prefer a median sternotomy and utilization of cardiopulmonary bypass; additional suggestions include the use of microvascular techniques and the creation of a wide, spatulated anastomosis. In our patient, restudy a year after operation confirmed wide patency of the reconstructed left pulmonary artery and normal pulmonary arterial flow distribution shown by lung scan. It is important to emphasize that, in addition to secondary tracheomalacia, congenital tracheal abnormalities can occur in association with the compression caused by pulmonary artery sling[16]; circumferential narrow tracheal cartilages have also been reported.[21] In such patients, relief of the compression may not improve lower airway constriction and a procedure on the trachea itself may be necessary. Many advances have been made in tracheal surgical procedures and we now believe that the distal half of the trachea can be resected with excellent expectations of success. Serial bronchoscopic evaluation is an integral part of the overall care of these patients.

Tetralogy of Fallot with an absent pulmonary valve is a rare condition which is quite difficult to manage.[17] Patients presenting in infancy usually have symptoms of airway compression due to aneurysmal dilatation of the pulmonary arteries; severe hypoxemia is uncommon in these infants. Few surgeons have extensive experience with this lesion. It is the general consensus that the usual repair of tetralogy (closure of the ventricular septal detect and enlargement of the right ventricular outflow tract) is not adequate therapy for this lesion. As a minimum, a new pulmonary valve should be implanted and the surgeon should consider transplantation of the right pulmonary artery to a position anterior to the superior vena cava. The postoperative course tends to be complicated. The right mainstem bronchus may cause troublesome airway narrowing for months after the operation. In successful cases the bronchial narrowing has improved and eventually the children are weaned from the ventilator and discharged from hospital.

In the modern era it is most unusual to see a patient with such enlargement of the left atrium that left mainstem bronchus compression is noted. Treatment is directed at correction of the intracardiac defect producing left atrial enlargement. Results can be excellent. Recent improvements in the development of bioprosthetic heart valves may prove useful when valve implantation is required in infancy.

Summary

DR. WALDMAN: *In vascular compression of the upper airway in children, symptoms tend to start early and be variable; growth is usually normal. Multiple radiographic studies are available for evaluating the airway, including high kilovoltage films, barium swallow, computerized axial tomographic* (CAT) *scanning and bronchography. Impairment of the cough and other respiratory defense mechanisms leads to recurrent infection, respiratory failure, or stridor and wheezing. Compression of the upper airway can be caused by three vascular structures: the aorta or its main branches, the pulmonary artery branches and the left atrium. Surgical therapy is available for essentially all forms of vascular compression but must be based on a detailed understanding of the anatomy and pathophysiology. The diagnostic*

and therapeutic approach must be individually determined in every case.

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The RH Factor in Emergency Transfusions

WE NORMALLY GIVE Rh-positive people Rh-positive blood, but you can give Rh-negative blood; and in an Rh-negative recipient, you can give Rh-positive blood if it is really a life-threatening circumstance. In a woman who may become pregnant in the future, one would not like to immunize her to Rh. But when you have a ruptured aortic aneurysm in a 55-year-old man or woman and you only have two or three units of Rh-negative blood, the thing to do is switch. You really are not going to cause any problems to that patient. They may become immunized to Rh some months later and they may, if they get future transfusions, have to get Rh-negative blood; but when it comes to a question of lifesaving, we are not too hesitant about switching the Rh-negative patient to Rh-positive blood.

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